

Ménétrier's Disease

National Digestive Diseases Information Clearinghouse



National Institute of
Diabetes and Digestive
and Kidney Diseases

What is Ménétrier's disease?

Ménétrier's disease causes the ridges along the inside of the stomach wall—called rugae—to enlarge, forming giant folds in the stomach lining. The rugae enlarge because of an overgrowth of mucous cells in the stomach wall.

In a normal stomach, mucous cells in the rugae release protein-containing mucus. The mucous cells in enlarged rugae release too much mucus, causing proteins to leak from the blood into the stomach. This shortage of protein in the blood is known as hypoproteinemia. Ménétrier's disease also reduces the number of acid-producing cells in the stomach, which decreases stomach acid.

Ménétrier's disease is also called Ménétrier disease or hypoproteinemic hypertrophic gastropathy.

What causes Ménétrier's disease?

Scientists are unsure about what causes Ménétrier's disease; however, researchers think that most people acquire, rather than inherit, the disease. In extremely rare cases, siblings have developed Ménétrier's disease as children, suggesting a genetic link.

Studies suggest that people with Ménétrier's disease have stomachs that make abnormally high amounts of a protein called transforming growth factor-alpha (TGF- α).

TGF- α binds to and activates a receptor called epidermal growth factor receptor. Growth factors are proteins in the body that tell cells what to do, such as grow larger, change shape, or divide to make more cells. Researchers have not yet found a cause for the overproduction of TGF- α .

Some studies have found cases of people with Ménétrier's disease who also had *Helicobacter pylori* (*H. pylori*) infection. *H. pylori* is a bacterium that is a cause of peptic ulcers, or sores on the lining of the stomach or the duodenum, the first part of the small intestine. In these cases, treatment for *H. pylori* reversed and improved the symptoms of Ménétrier's disease.¹

Researchers have linked some cases of Ménétrier's disease in children to infection with cytomegalovirus (CMV). CMV is one of the herpes viruses. This group of viruses includes the herpes simplex viruses, which cause chickenpox, shingles, and infectious mononucleosis, also known as mono. Most healthy children and adults infected with CMV have no symptoms and may not even know they have an infection. However, in people with a weakened immune system, CMV can cause serious disease, such as retinitis, which can lead to blindness.

Researchers are not sure how *H. pylori* and CMV infections contribute to the development of Ménétrier's disease.

¹Fretzayas A, Moustaki M, Alexopoulou E, Nicolaidou P. Ménétrier's disease associated with *Helicobacter pylori*: three cases with sonographic findings and a literature review. *Annals of Tropical Paediatrics*. 2011;31(2):141–147.

Who gets Ménétrier's disease?

Ménétrier's disease is rare. The disease is more common in men than in women. The average age at diagnosis is 55.²

What are the signs and symptoms of Ménétrier's disease?

The most common symptom of Ménétrier's disease is pain in the upper middle part of the abdomen. The abdomen is the area between the chest and hips.

Other signs and symptoms of Ménétrier's disease may include

- nausea and frequent vomiting
- diarrhea
- loss of appetite
- extreme weight loss
- malnutrition
- low levels of protein in the blood
- swelling of the face, abdomen, limbs, and feet due to low levels of protein in the blood
- anemia—too few red blood cells in the body, which prevents the body from getting enough oxygen—due to bleeding in the stomach

People with Ménétrier's disease have a higher chance of developing stomach cancer, also called gastric cancer.

How is Ménétrier's disease diagnosed?

Health care providers base the diagnosis of Ménétrier's disease on a combination of symptoms, lab findings, findings on upper gastrointestinal (GI) endoscopy, and stomach biopsy results. A health care provider will begin the diagnosis of Ménétrier's disease by taking a patient's medical and family history and performing a physical exam. However, a health care provider will confirm the diagnosis of Ménétrier's disease through a computerized tomography (CT) scan, an upper GI endoscopy, and a biopsy of stomach tissue. A health care provider also may order blood tests to check for infection with *H. pylori* or CMV.

Medical and family history. Taking a medical and family history is one of the first things a health care provider may do to help diagnose Ménétrier's disease. He or she will ask the patient to provide a medical and family history.

Physical exam. A physical exam may help diagnose Ménétrier's disease. During a physical exam, a health care provider usually

- examines a patient's body
- uses a stethoscope to listen to bodily sounds
- taps on specific areas of the patient's body

²Coffey R, Washington M, Corless C, Heinrich M. Ménétrier disease and gastrointestinal stromal tumors: hyperproliferative disorders of the stomach. *The Journal of Clinical Investigation*. 2007;117(1):70–80.

CT scan. CT scans use a combination of x rays and computer technology to create images. For a CT scan, a health care provider may give the patient a solution to drink and an injection of a special dye, called contrast medium. CT scans require the patient to lie on a table that slides into a tunnel-shaped device where an x-ray technician takes x rays. An x-ray technician performs the procedure in an outpatient center or a hospital, and a radiologist—a doctor who specializes in medical imaging—interprets them. The patient does not need anesthesia. CT scans can show enlarged folds in the stomach wall.

Upper GI endoscopy. This procedure involves using an endoscope—a small, flexible tube with a light—to see the upper GI tract, which includes the esophagus, stomach, and duodenum. A gastroenterologist—a doctor who specializes in digestive diseases—performs the test at a hospital or an outpatient center. The gastroenterologist carefully feeds the endoscope down the esophagus and into the stomach. A small camera mounted on the endoscope transmits a video image to a monitor, allowing close examination of the stomach lining. The gastroenterologist also can take a biopsy of the stomach tissue during the endoscopy. A health care provider may give a patient a liquid anesthetic to gargle or may spray anesthetic on the back of the patient's throat. A health care provider will place an intravenous (IV) needle in a vein in the arm to administer sedation. Sedatives help patients stay relaxed and comfortable. The test can show enlarged folds in the stomach wall.

Biopsy. Biopsy is a procedure that involves taking a piece of stomach tissue for examination with a microscope. A gastroenterologist performs the biopsy at the time of upper GI endoscopy. A pathologist—a doctor who specializes in diagnosing diseases—examines the stomach tissue in a lab. The test can diagnose Ménétrier's disease by showing changes in the stomach's mucous cells and acid-producing cells.

Blood test. A health care provider will take a blood sample that can show the presence of infection with *H. pylori* or CMV. A blood test involves drawing blood at a health care provider's office or a commercial facility and sending the sample to a lab for analysis.

How is Ménétrier's disease treated?

Treatment may include medications, IV protein, blood transfusions, and surgery.

Medications

Health care providers may prescribe the anticancer medication cetuximab (Erbix) to treat Ménétrier's disease. Studies have shown that cetuximab blocks the activity of epidermal growth factor receptor and can significantly improve a person's symptoms, as well as decrease the thickness of the stomach wall from the overgrowth of mucous cells. A person receives cetuximab by IV in a health care provider's office or an outpatient center. Studies to assess the effectiveness of cetuximab to treat Ménétrier's disease are ongoing. A health care provider also may prescribe medications to relieve nausea and abdominal pain.

In people with Ménétrier's disease who also have *H. pylori* or CMV infection, treatment of the infection may improve symptoms. Health care providers prescribe antibiotics to kill *H. pylori*. Antibiotic regimens may differ throughout the world because some strains of *H. pylori* have become resistant to certain antibiotics—meaning that an antibiotic that once destroyed the bacterium is no longer effective. Health care providers use antiviral medications to treat CMV infection in a person with a weakened immune system in order to prevent a serious disease from developing as a result of CMV. Antiviral medications cannot kill CMV; however, they can slow down the virus reproduction.

Intravenous Protein and Blood Transfusions

A health care provider may recommend an IV treatment of protein and a blood transfusion to a person who is malnourished or anemic because of Ménétrier's disease. In most cases of children with Ménétrier's disease who also have had CMV infection, treatment with protein and a blood transfusion led to a full recovery.

Surgery

If a person has severe Ménétrier's disease with significant protein loss, a surgeon may need to remove part or all of the stomach in a surgery called gastrectomy.

Surgeons perform gastrectomy in a hospital. The patient will require general anesthesia. Some surgeons perform a gastrectomy through laparoscopic surgery rather than through a wide incision in the abdomen. In laparoscopic surgery, the surgeon uses several smaller incisions and feeds special surgical tools through the incisions to remove the diseased part of the stomach. After gastrectomy, the surgeon may reconstruct the changed portions of the GI tract so that it may continue to function. Usually the surgeon attaches the small intestine to any remaining portion of the stomach or to the esophagus if he or she removed the entire stomach.

Eating, Diet, and Nutrition

Researchers have not found that eating, diet, and nutrition play a role in causing or preventing Ménétrier's disease. In some cases, a health care provider may prescribe a high-protein diet to offset the loss of protein due to Ménétrier's disease. Some people with severe malnutrition may require IV nutrition, which is called total parenteral nutrition (TPN). TPN is a method of providing an IV liquid food mixture through a special tube in the chest.

Points to Remember

- Ménétrier's disease causes the ridges along the inside of the stomach wall—called rugae—to enlarge, forming giant folds in the stomach lining. The rugae enlarge because of an overgrowth of mucous cells in the stomach wall.
- Scientists are unsure about what causes Ménétrier's disease; however, researchers think that most people acquire, rather than inherit, the disease.
- Ménétrier's disease is rare. The disease is more common in men than in women.
- The most common symptom of Ménétrier's disease is pain in the upper middle part of the abdomen.
- Health care providers base the diagnosis of Ménétrier's disease on a combination of symptoms, lab findings, findings on upper gastrointestinal (GI) endoscopy, and stomach biopsy results.
- Treatment may include medications, intravenous (IV) protein, blood transfusions, and surgery.

Hope through Research

The National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK) conducts and supports basic and clinical research into many digestive disorders, including Ménétrier's disease.

Clinical trials are research studies involving people. Clinical trials look at safe and effective new ways to prevent, detect, or treat disease. Researchers also use clinical trials to look at other aspects of care, such as improving the quality of life for people with chronic illnesses. To learn more about clinical trials, why they matter, and how to participate, visit the NIH Clinical Research Trials and You website at www.nih.gov/health/clinicaltrials. For information about current studies, visit www.ClinicalTrials.gov.

For More Information

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